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Multiple Widespread Blue Nodules: A Clinicopathological Case

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Key Words

Glomangiomas · Glomus tumours · Blue nodules

Abstract

Glomangiomas are a variant of benign glomus tumours; glomus tumours are benign neoplastic proliferations of the glomus body, which is made up of perivascular smooth muscle cells. This forms arteriovenous anastomoses which play an important role in vascular regulation of skin temperature. The aim of this article is to describe 2 cases of hereditary glomangiomas.

Case Reports

The first case is a 29-year-old female who presented with a 2-year history of multiple bluish nodules particularly on her trunk and limbs, ranging from 0.5–2.5 cm in size. Although some of the lesions had been present since she was 5 years old, their accelerating growth over the past year had caused concern. There were no other systemic features present. Her mother also reported 2–3 similar lesions, which had developed during adolescence, on her upper limbs but no investigation had been conducted in the past. No other family member was affected. Several soft, partially compressible nontender blue, nodular and multifocal lesions ranging from 0.5 to 2.5 cm in diameter were noted on the back, flanks, thighs, upper arms, palms and soles ([fig. 1a, b](#)). Most of the lesions were discrete but there were a few that coalesced.

The second case is a 14-year-old female who presented with multiple subcutaneous nodules on the leg and the trunk, some of which were painful. Her younger sister, aged 6 years old, was noted to have similar lesions on her feet ([fig. 1c](#)). Her mother, aged 49 years old, had similar lesions on her trunk, especially on the back and across the shoulder.

Excision biopsy taken from both case 1 (nodules in her lower back) and case 2 (right leg) showed a nonencapsulated intradermal tumour composed of thin-walled vascular channels lined by endothelial cells. These were surrounded by irregular layers of cuboidal cells, in keeping with glomus-like cells ([fig. 2a, b](#)), which stained positively for smooth muscle actin on immunohistochemistry ([fig. 2c](#)). Staining for S-100 was negative. The diagnosis was hereditary glomangiomas in both cases.

Discussion

Glomangiomas are a variant of benign glomus tumours. Glomus tumours are benign neoplastic proliferations of the glomus body, which is made up of perivascular smooth muscle cells and forms arteriovenous anastomoses, which play an important role in vascular regulation of skin temperature [1]. The occurrence of malignant glomus tumours is exceedingly rare [2].

Glomus tumours are characteristically solitary benign lesions. They present in young individuals as small papules or nodules and are blue or bluish-purple in appearance. The most commonly affected sites are the distal extremities, such as the nail bed or the palm. Glomus tumours are typically painful and tender to touch, especially in response to changes in temperature or pressure. Histologically, there are solid sheets of glomus cells around small blood vessels [3, 4].

Glomangiomas differ clinically from glomus tumours in that they occur in childhood and adolescence, do not have a predilection for the subungal region, and often are multifocal. Rarely can they be congenital. Furthermore, they are often painless although they may be tender to touch. They are usually multiple, soft, red-to-blue nodules but may appear as pink-to-deep-blue plaque-like lesions. They are often widespread, and can join together to become larger plaques. They tend to get thicker and bluer with age.

The histopathology of glomangiomas differs from glomus tumours in that there are dilated venous channels, resembling venous malformations which stain positively for α -smooth muscle actin. They are less circumscribed than glomus tumours [5].

Most glomangiomas manifest sporadically; however, there have been reports showing autosomal-dominant inheritance patterns. Our cases, both having a strong familiarity with many members affected on different generations, all seem to be of familial type, with dominant inheritance. The chromosome 1p21–p22 is thought to be involved, and it has been suggested that this results in loss of function of the protein glomulin, which is considered important for smooth muscle cell differentiation [6, 7].

Due to the benign, indolent course, often no treatment is required. However, surgical excision can be used for symptomatic lesions. Excision may be more difficult for multiple glomus tumours because of their poor circumscription and the large number of lesions. For these cases, there is a range of possible treatments, including sclerotherapy, electron-beam radiation, argon and CO₂ lasers or observation of asymptomatic lesions. The prognosis for excised glomus tumours is very good, with a low recurrence rate [8].

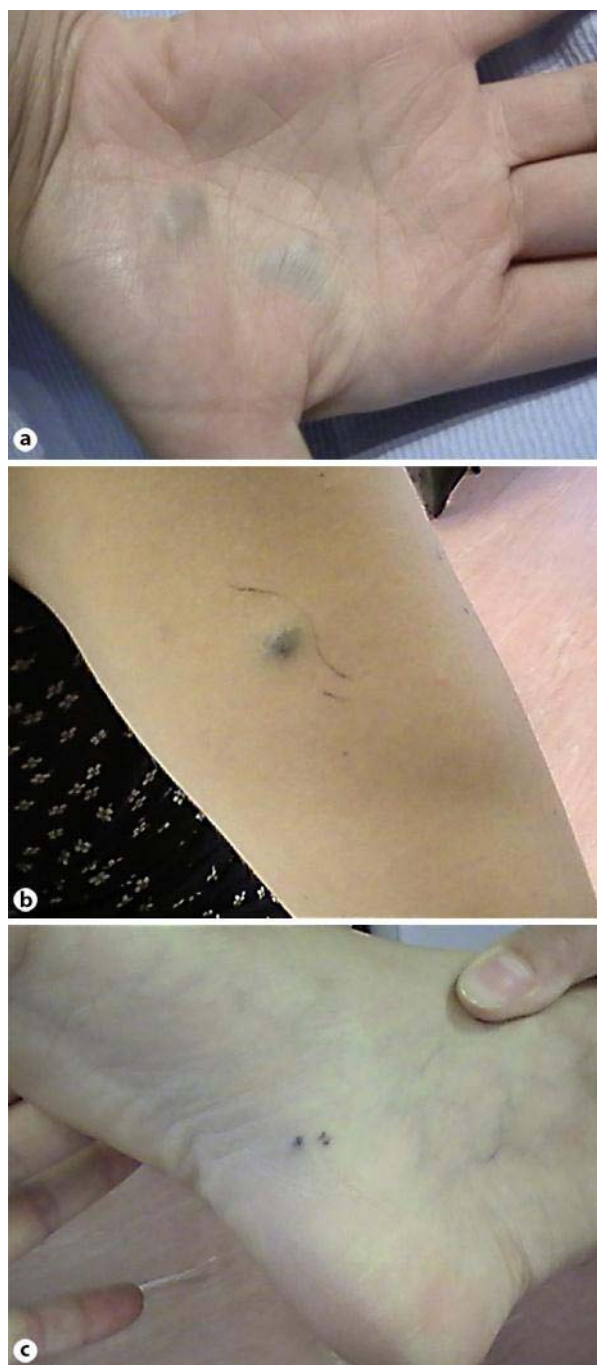


Fig. 1. Blue nodules **a** on the palm of the hand, **b** on the elbow, and **c** on the foot.

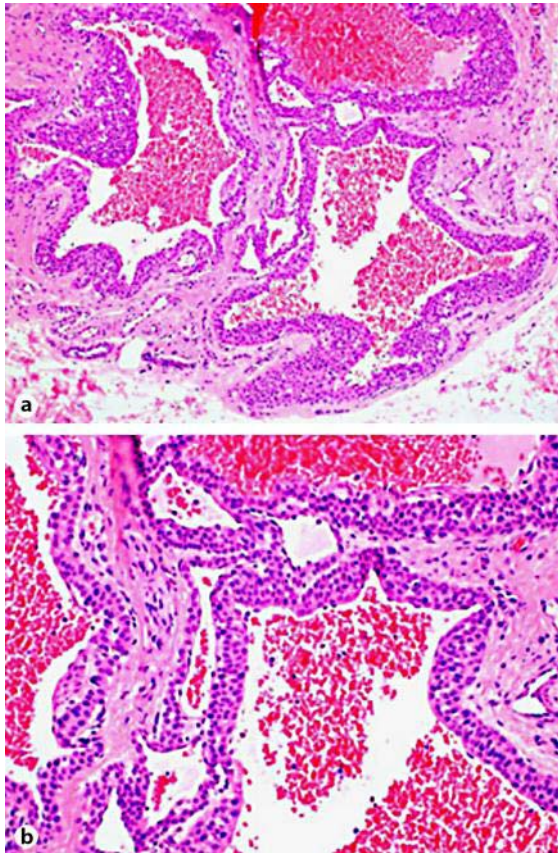


Fig. 2. **a** Intradermal thin-walled folded and dilated vascular channels (haematoxylin and eosin, original magnification $\times 50$). **b** Intradermal vascular channels lined by benign flat endothelial cells and monomorphic round glomus-like cells with large nuclei and slightly eosinophilic cytoplasm (haematoxylin and eosin, original magnification $\times 350$).

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